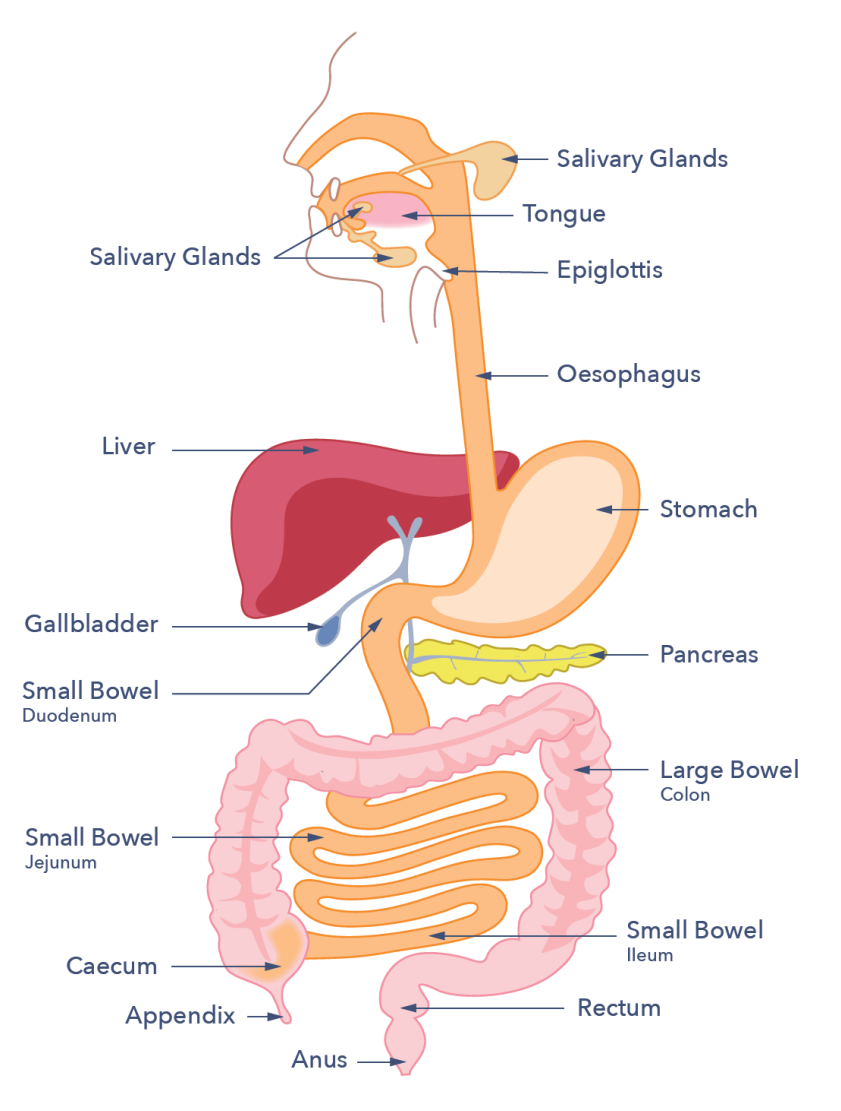




## FUNDING RESEARCH TO FIGHT DISEASES OF THE GUT, LIVER & PANCREAS



## **THIS FACTSHEET IS ABOUT CHRONIC INTESTINAL PSEUDO-OBSTRUCTION**

Chronic Intestinal Pseudo-obstruction (CIPO) is an overall term for several rare conditions which can affect any part of the gut (intestine) but primarily the small intestine. Symptoms resemble those caused by a blockage (obstruction) of the gut, but when investigated no blockage is found (hence the term “pseudo” which means false). CIPO is caused by an abnormality in the nerves or muscles of the gut, which leads to altered and inefficient contractions (peristalsis) of the digestive tract, so gut content does not pass through properly. When a specific abnormality of the intestinal nerves or muscles is identified, the diagnosis of CIPO may be refined to include this. Terms such as visceral myopathy (bowel muscle dysfunction), visceral neuropathy (bowel nerve dysfunction) and mesenteric plexitis (inflammation of part of the nervous system of the gut) could be used. All fall within the umbrella of causes of CIPO.

Numbers of people who live with CIPO are not well established but in Japanese adults there were reported to be 9 people in 1000,000<sup>1</sup>, which is defined as rare. There are slightly higher numbers of males than females diagnosed.

### **CAUSES**

#### **CAUSES OF CHRONIC INTESTINAL PSEUDO-OBSTRUCTION**

When food is ingested, the gut usually has co-ordinated muscle contractions to help it pass through. However, in CIPO this does not happen, the contractions are weak or uncoordinated and gut content does not pass through properly. In most cases, what has caused this damage and weakness to the muscle and/or nerves of the gut is not clear. Occasionally, CIPO can be a result of another disease not related to the gut, for example autoimmune conditions such as lupus and rheumatoid arthritis, disorders affecting hormone regulation such as diabetes, underactive thyroid or neurological disorders such as Parkinson’s disease and also diseases affecting the connective tissue such as systemic sclerosis. It can also, rarely, be caused by infections such as herpes viruses, or following abdominal surgery and also affect other areas of the digestive tract.

Some forms of cancer can cause CIPO and, more rarely, it is caused by certain medications such as opioids (including codeine, tramadol, morphine, buprenorphine, tapentadol and fentanyl). Do not stop taking any medications without discussing it with your doctor. In some cases, it has a genetic component and runs in families. The condition can affect people at any age including rarely infants and children, where the cause often runs in families.

#### **HOW IS CHRONIC INTESTINAL PSEUDO-OBSTRUCTION DIAGNOSED?**

Investigations may include:

**Radiology tests: including abdominal X-ray, CT scan, barium small bowel follow through and MR small bowel.** These will help to rule out any mechanical obstruction in the bowel and will typically show an enlarged (“dilated”) intestine. If these scans do

not reveal anything unusual then transit studies of the gut may be performed to see how content travels through the gut.

**Manometry (pressure studies):** these are, rarely, used to measure contractions of the gut.

**Blood tests:** these are needed to look for anaemia, infections, hormone levels and antibodies and will also help to establish whether a cause can be found for CIPO.

**Biopsy:** if the results of the above tests are normal, some people may go on to have a biopsy (tissue sample) of the gut, so the muscles and nerves can be directly visualised and examined under a microscope. However, this usually involves surgery, which in itself has risks and rarely affects treatment, so is not frequently undertaken.

It is important to note that although there are various methods of investigating for causes of CIPO, in many cases all the results (except for the dilated bowel) will be normal, meaning an exact cause of CIPO is often not found which is called “idiopathic”. In many cases the diagnosis is made only after a long delay and despite many unhelpful investigations.

## HOW CAN CHRONIC INTESTINAL PSEUDO-OBSTRUCTION AFFECT YOU OVER TIME?

For some people with CIPO, medications alone may be helpful. However, in general, people with CIPO tend to have ongoing symptoms, despite treatment. Sadly, in some cases the condition can be progressive and deteriorates with time.

### SYMPTOMS

CIPO can impact on your life via the symptoms it causes such as pain, nausea and vomiting as well as their effects on your general wellbeing due to weight loss and malnutrition. You may also experience bloating and distension [add link], feel full very quickly and experience symptoms of heartburn and reflux [add link]. This might also be accompanied by a change in bowel function such as constipation and/or diarrhoea [add links].

**Quality of life:** CIPO can be a long-term condition with symptoms that occur frequently or are constant. This can affect daily activities including family, work and social life. It is recognised that recurrent severe symptoms with no clear diagnosis can cause major upset and feelings of hopelessness amongst many patients. If you are experiencing any of these feelings it is important to let the doctor know, so that appropriate support can be organised.

### TREATMENT

#### WHAT TREATMENT IS AVAILABLE?

The treatment of CIPO is usually individual and can be challenging. You may need treatment or support from many different specialists including a gastroenterologist (gut doctor), a dietitian, pain management team, a psychologist and, occasionally, a surgeon. These team members often need to work together to ensure an individualised treatment plan, according to symptoms, age and personal preference. Treatment can

include diet changes, nutritional support, medications, decompression of the gut or surgery. If CIPO is due to another condition, it is important that the underlying disorder is treated.

**Diet and nutritional support:** some people with CIPO may become malnourished. This can be potentially life-threatening so it is important that a dietician is involved at an early stage and can advise on certain diets, depending on individual needs. People with CIPO are generally advised to eat small meals and often, rather than three large meals a day. Whilst there is little evidence for food intolerances in CIPO it is best to work with a dietitian specialising in gastroenterology who will support you to find the best diet for you so ask your doctor for a referral. If adequate nutrition cannot be maintained through eating food, then other feeding methods may be needed such as NG (nasogastric) or PEG (percutaneous endoscopic gastrostomy) feeding or PN (parenteral nutrition) where feed is delivered directly into a vein. Parenteral nutrition is considered only in severe cases as it can also have some severe side effects as a treatment.

**Medications:** medications are used to help treat symptoms associated with CIPO but do not cure it. Prokinetic medicines (drugs that increase movement) which aim to improve gut movement by increasing the frequency of contractions can be used. Some examples include domperidone, metoclopramide, erythromycin, octreotide and prucalopride. For nausea, anti-sickness medications can be used. If pain is severe, painkillers can be prescribed. However, it is important to be aware that morphine and morphine derivatives (such as codeine) can make symptoms worse, should generally be avoided and may need to be stopped if already started. Low dose anti-depressants and gabapentin can be used to treat chronic pain. Small intestinal bacterial overgrowth is a common complication of CIPO and may require antibiotic therapy.

**Decompression:** if the gut becomes very large and the patient does not respond to medications, the excess gas may need to be removed in a procedure called decompression. This helps reduce the pressure in the digestive tract. It can be performed using tubes passed into the stomach (through the mouth or nose) or into the bowel (through the rectum, back passage). Some patients gain relief from a large-calibre PEG tube known as a venting PEG to allow wind (and sometimes fluid) to pass out from the stomach.

**Surgery:** this is reserved for severe cases and is rarely performed as CIPO normally affects the whole gut. However, if CIPO is thought to be limited to one section of the gut, surgery may be used to form a venting stoma (artificial opening to reduce pressure) or remove that portion. Extremely rarely, patients may be considered for intestinal transplantation. In general, a key goal is to avoid surgical intervention where possible

## **DOES CHRONIC-INTESTINAL PSEUDO OBSTRUCTION NEED MONITORING AND IF SO HOW?**

If the symptoms of CIPO are well controlled, then regular follow-up with a doctor is not necessary. However, if symptoms are ongoing, regular follow-up is useful to ensure that new treatments for symptom control can be considered and nutrition is monitored. If nutritional support is required, then regular follow-up with a doctor, dietitian and/or nutrition nurse is important to monitor weight and ensure the feeding regime is adequate. If you have a tube feed this would be monitored in the community, and you will be provided contact details in case of any problems.

## **SUPPORT**

### **WHAT TO ASK YOUR DOCTOR?**

These are some useful questions you can ask your doctor:

- Has CIPO been considered and thoroughly investigated?
- May I be referred to a dietitian to see if there are any changes to my diet or support to stop my weight loss?
- What form of pain relief should I take – at home to prevent attacks and in hospital when I have an attack?
- How often do I need follow-up?

### **WHERE CAN I GET MORE INFORMATION?**

More information is available from the charity **PORT** who support people who have been diagnosed with CIPO.

<http://www.port-charity.org.uk/>

The British society of Gastroenterology have produced guidelines which cover causes, diagnosis and management of CIPO in detail.

<https://www.bsg.org.uk/clinical-resource/the-management-of-adult-patients-with-severe-chronic-small-intestinal-dysmotility/>

### **RESEARCH**

Unfortunately, there is no cure for CIPO. For most patients there are treatments that provide some relief, but this is often not complete. CIPO is relatively rare and has not received much research interest or funding. Further research into understanding the cause of symptoms is urgently needed to help in the development of new and better treatments. More specific medications or bowel pacemakers, which help to maintain the muscle contractions in the gut, could in the future help to manage symptoms more effectively.

**[For further information, visit gutscharity.org.uk](http://gutscharity.org.uk)**